Osteochondroses

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Abstract

Osteochondrosis is an umbrella term for a broad variety of conditions affecting all areas of the body. The aetiology is not fully understood and the clinical effects vary from mild pain and discomfort to gross disability. Generally they are self-limiting conditions and can be managed with activity modification and physical therapy. However, some conditions, such as Legg–Calve–Perthes, or Blount's, can result in lifelong disability if left untreated.

This paper aims to give a broad overview of the osteochondroses and present a synopsis of the more common types, reviewing the current literature and giving guidance on management.

Keywords children; deformity; osteochondrosis; osteonecrosis

Introduction

"Osteochondroses" is a collective term for a heterogeneous group of conditions characterized by an abnormality in endochondral ossification in a previously normal skeleton.[1] This group encompasses abnormalities within articular cartilage, physes, tendon and ligament insertions and other impact sites. Together they represent a spectrum from sub-clinical to severely disabling with different aetiologies. There are over 50 types described in the literature, often with eponymous names and they can occur at separate sites within the body, or indeed distinct osteochondroses can occur at synchronous sites within the same individual. Examples of common eponymous conditions are seen in Figure 1.

A degree of confusion surrounds this broad subject in the literature, as the nomenclature is often used inaccurately and interchangeably. Originally thought to have a significant inflammatory aetiology, Konig in 1887 coined the term "Osteochondritis". The inflammatory component has been in dispute throughout the latter half of the 20th century. The true aetiology remains unknown. A genetic predisposition has been linked to several specific osteochondroses. Confusingly, the postfixes commonly used for specific osteochondroses are also used interchangeably; disease, syndrome and phenomena can be seen throughout the literature.

Due to the breadth of conditions covered by this broad umbrella term, this article aims to cover the common osteochondroses, highlighting the specific clinical and pathological features, with current strategies of management.

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Pathophysiology

A unifying primary pathophysiological process is yet to be identified, however all of the osteochondroses follow the same sequence of events: a vascular event resulting in ischaemia and necrosis of the bone and cartilage, with secondary changes such as fragmentation, collapse, and remodelling. Histologically, the cascade following ischaemic necrosis resulting in revascularization, formation of granulation tissue, osteoclastic resorption and osteoblastic deposition of new bone supports this above common process.

Proposed hypotheses into the primary vascular event relate to a genetic predisposition to a hypercoaguable state, acute or repetitive micro-trauma, infection, or congenital malformations. Interestingly, when this phenomenon affects the epiphysis in a joint, it is usually the convex joint surface which is involved and not the concave side, supporting the concept that the compressive impact of repetitive trauma in the adolescent age could be one of the causes. However, at an apophysis, where a tendon or ligament is attached to the growth plate, it is under tension or traction and referred to as an apophysitis.

Classification

Several classifications have been used historically. Outdated systems such as Burrow's and Goff's subclassify by pressure, traction and atavistic thus inaccurately classifying by aetiology. The widely accepted use of Siffert's classification (1981) classifies by anatomical region.[1]

Articular osteochondroses

This subgroup causes irregularity to the articular surfaces of synovial joints and may be primary or secondary. Primary articular osteochondroses include Freiberg's disease (occurring at the metatarsal head), or Panner's disease (occurring at the capitellum). Secondary articular osteochondroses are a group of conditions affecting the bone adjacent to the articular surface which indirectly affects the joint surface such as Legg–Calve–Perthes disease.

Non-articular osteochondroses

Typified by pain and swelling at entheses, this group of conditions is often related to activity and considered to be an overuse phenomenon. They are self-limiting and generally managed conservatively. They can be further sub-classified into those which affect tendon insertions (Osgood–Schlatter syndrome affecting the insertion of the patellar tendon, and Monde–Felix disease affecting the proximal femur), ligament insertions (Adam's disease affecting the medial epicondyle of the elbow, and vertebral ring apophysis), or impact sites (Sever's disease affecting the calcaneum).

Physeal osteochondroses

Any condition affecting the physes can result in growth abnormality, deformity or limb length discrepancy, and as a result these often require treatment. They can be sub-classified into conditions which affect the axial and appendicular skeleton. Axial conditions include Scheuermann's disease resulting in pathological thoracic kyphosis, Appendicular conditions include

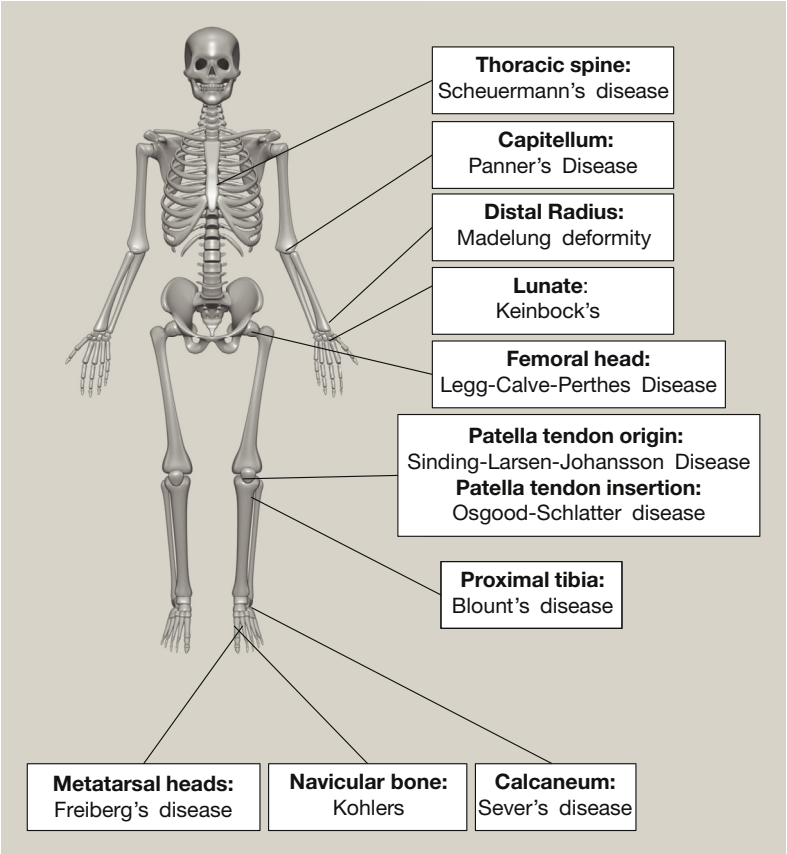

Figure 1 Anatomical sites of common osteochondrosis.

Blount's disease causing excessive tibia vara, or Madelung's deformity affecting the distal radial epiphysis.

Clinical assessment

Osteochondroses have been reported to constitute up to 10% of presentations of musculoskeletal pain to Primary Care and are often overlooked.[2] There is often an indolent onset and non-specific examination findings such as localized tenderness, limited range of motion and swelling. The diagnosis is often confirmed late after the onset of disability.

Clinicians should be vigilant when musculoskeletal pathology presents alongside systemic upset and these patients should be investigated further to exclude alternative diagnoses such as infection or malignancy. Night pain is a red flag symptom for the latter two conditions.

Appropriate imaging alongside clinical assessment should confirm the diagnosis. Plain radiographs are helpful but often the changes of osteochondroses develop later. Magnetic resonance imaging (MRI) is more sensitive and can show subtle features of ischaemia/revascularization.

Articular osteochondrosis

Freiberg's disease

Freiberg's is an osteochondrosis affecting the metatarsal heads. Typically, this affects the second metatarsal, rarely affecting the other metatarsals, in adolescent girls who participate in regular activities such as ballet dancing.[3] Freiberg himself postulated that an abnormally long metatarsal may contribute, but alongside several other authors also suggests that it may be a vascular phenomenon. The true aetiology remains unknown and alongside repetitive trauma it is likely to be multifactorial. Resultant ischaemia and possible fracture through the epiphysis occurs and can give rise to chronic pain in the forefoot. Bilateral disease occurs in up to 10% of sufferers.[4] Pathologically, a delay to union with poor endochondral ossification can result in a misshapen metatarso-phalangeal joint and ultimately, if untreated, a rigid painful osteoarthritic joint. Patients present with poorly localized forefoot pain which is related to activity. Confirmatory clinical findings may be sparse. Tenderness around the metatarsal head may be the only positive finding.

As with most osteochondroses, plain radiographs should be the first line of investigation and these are sometimes normal, or sometimes show very subtle changes in the early stages. Magnetic resonance imaging delineates the more discrete changes. Based on plain radiographs, Smillie described five stages of Freiberg's disease. Stage 1 is a fissure-type fracture through the epiphysis. Stage 2 is an abnormal contour of the articular surface with central bone resorption. Stage 3 shows progression of central bone resorption. Stage 4 has fragmentation and development of loose bodies. Stage 5 is complete flattening of the metatarsal head (Figure 2).[5]

Treatment goals are to allow resolution of pain, return normal function and prevent deformation of the metatarsal